Madigan Army Medical Center
Referral Guidelines

Neural Tube Defect

Diagnosis/Definition

- Disturbances in the development and closure of the neural tube includes a spectrum of anomalies ranging from defects of the cephalad neural tube (anencephaly) to occult spina bifida.
- Collectively, this spectrum of defects is variously referred to as meningocele, myelomeningocele, spina bifida, or myelodysplasia.

Initial Diagnosis and Management

The diagnosis of neural tube defect (NTD) is often made during prenatal screening. The diagnosis can also be made at birth with the appearance of the structural abnormality somewhere along the spinal axis. Surgical interventions, to include defect closure and shunt placement, will be determined and completed by the appropriate surgical specialists. Primary care providers will recognize and monitor the multisystem involvement of these patients: hydrocephalus, cranial nerve abnormalities, respiratory regulation dysfunction, urinary tract infection and dysfunction, and orthopedic deformities. A careful documentation of the level of motor and sensory nerve involvement and recognizing asymmetry in this involvement is important for future care and monitoring.

Ongoing Management and Objectives

- The goal of comprehensive treatment for these children is to achieve maximal levels of motor, intellectual and social functioning. This is achieved through a thorough understanding of the neurologic dysfunction, the associated medical problems, the level of cognitive function and the psychosocial adjustment.
- Objectives for management are to prevent or reduce deformities, to train the child in adaptive functioning and self-care skills, to achieve some means of independent locomotion, to control excretion of urine and feces, to foster personal adjustment and to provide proper education and vocational rehabilitation.
- Particular areas of concern will be shunt malfunction or infection, urinary tract infection, changes in urine or bowel patterns, changes in motor or sensory deficit levels, decubitus formation and orthopedic deformities.

Indications for Specialty Care Referral

All children initially suspected of having NTD should be referred to the Developmental Behavioral Pediatrics (DBP) Division for multidisciplinary evaluation in the Spina Bifida Clinic/Neuromuscular Clinic (includes DBP, Pediatric Neurology, Pediatric PT, Orthopedics, and Urology). Children with an established diagnosis of NTD may also be referred for review of current needs to include medical care, equipment, and educational services/case management on an every 6-12 month basis. Further consultation may also be required when there is a deviation from the expected clinical course or new findings or complications that are associated with NTD are identified. All referrals for NTD should be reviewed by the Developmental Behavioral Pediatrics clinic prior to release to outside agencies.

Criteria for Return to Primary Care

- Completed specialty care evaluation with established diagnosis, functional levels and recommendations that can be accomplished at a primary care level.
- A level of NTD involvement that can be managed with primary care and ongoing monitoring by specialists (the majority of NTD patients).

References

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Shaer CM, Chescheir N, Schulkin J. Myelomeningocele: A review of the epidemiology, genetics, risk factors for conception, prenatal diagnosis, and prognosis for affected individuals. Obstetrical and Gynecological Survey. 62:7, 2007

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Referral Guidelines require review every three years.

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